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Gall-bladder polyps in Peutz-Jeghers syndrome

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Summary

A case of Peutz-Jeghers syndrome with polyps in the gall bladder is presented. No previous description of this finding has been reported. The literature regarding known sites of polyposis in this condition is reviewed.

Introduction

Peutz-Jeghers syndrome (PJS) is characterized by: (i) autosomal dominant inheritance; (ii) cutaneous pigmentation; (iii) gastro-intestinal polyposis. In all, more than 300 cases have been described with a world-wide distribution and no racial predilection.

In 1921 Peutz described 7 cases of multiple intestinal polyps associated with melanin spots on the lips, buccal mucosa and digits. Three generations of a Dutch family were affected. In 1949, Jeghers, McKusick and Katz described 10 further cases exhibiting the typical features and noted the familial incidence. Foster (1944) described a man and his daughter, both of whom had intussusception due to polyps, accompanied by oral pigmentation. The patient described below is the above daughter who 22 years later was found to have gall-bladder polyposis.

Case report

A 38-year-old woman was admitted with a 4-month history of biliary colic. She was known to have PJS, having undergone resection of an ileo-ileal intussusception in 1943. Her father and cousin were also known to have PJS.

Physical examination showed typical peri-oral pigmentation (Fig. 1). There was right hypochondrial tenderness with a positive Murphy's sign. Haematological and biochemical investigations were normal. Oral cholecystography demonstrated a large polypoidal defect in the gall bladder with several smaller defects thought to represent small polyps or stones (Fig. 2). At laparotomy the gall bladder was thickened and a soft mass was palpable within its lumen. The common bile duct was normal and operative cholangiography via the cystic duct showed no abnormality. Cholecystectomy was performed. Multiple lobulated polypi were present in the gall bladder with several small pigment stones (Fig. 3). Examination of the small bowel disclosed 2 polyps in the ileum and these were removed (Fig. 4). The patient made an uneventful postoperative recovery. Histological examination of the gall bladder showed several well differentiated adenomatous polyps. The small bowel polyps were non-proliferating hamartomas, typical of PJS.

Since 1965 the patient has remained well apart from a transient episode of intussusception in 1970 which responded to conservative treatment.

Discussion

Patients with PJS usually present with gastrointestinal symptoms of intussusception. Recurrent episodes of colicky abdominal pain usually commence in adolescence and there are often long symptom-free periods. Intussusceptions are frequently transient and self-limiting, but the occurrence of vomiting and abdominal distension indicates probable intestinal obstruction. Rectal bleeding or melaena is the second most frequent presentation. Less common symptoms include those related to prolapse of a rectal polyp or those due to presence of polyps in the urinary or respiratory tracts. Investigation of relatives of an affected patient may reveal asymptomatic cases of PJS.

Polyps in PJS are typically multiple. They occur mainly in the gastrointestinal tract, although adenomas have been described in the ureter (Sommerhaug and Mason, 1970) and in the bladder, renal pelvis, bronchus and nasal passages (Dormandy, 1957; Dodds *et al.*, 1971). The small bowel is involved in more than 95% of patients, the colon in approximately 30% and the stomach in about 25%. Polyps may arise in the appendix and rarely in the oesophagus (André, Duhamel and Bruaire, 1966). The present case represents the first report of gall bladder polyposis in PJS.

The relationship between PJS and gastrointestinal

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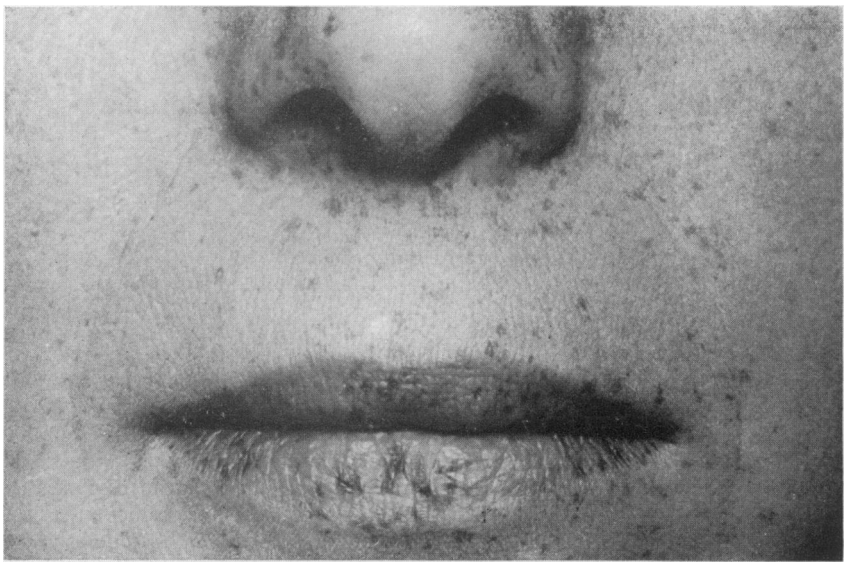

FIG. 1. Peri-oral pigmentation.

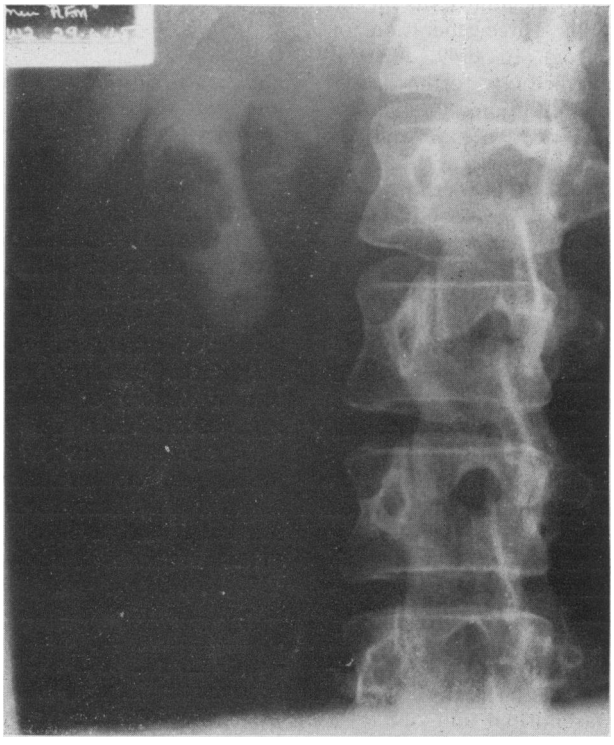

FIG. 2. Oral cholecystogram, showing large polypoidal filling defect with multiple small calculi.

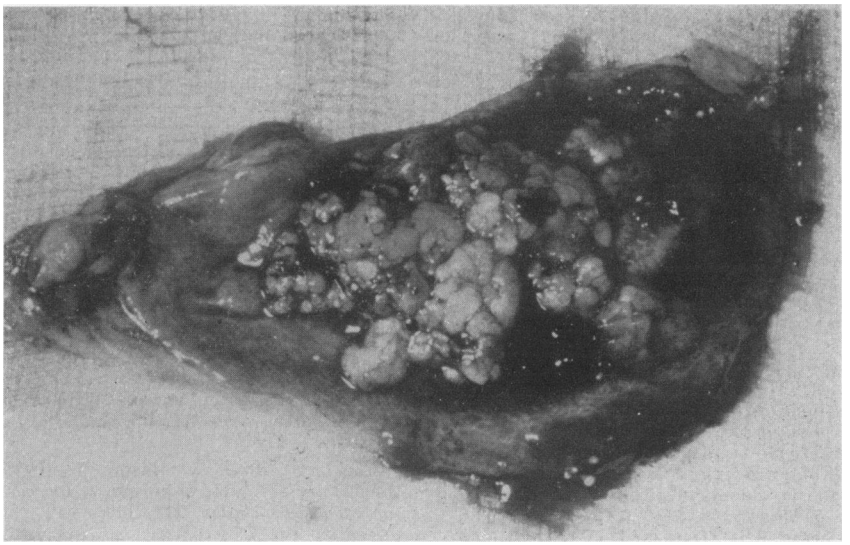

FIG. 3. Excised gallbladder with multiple lobulated polyps.

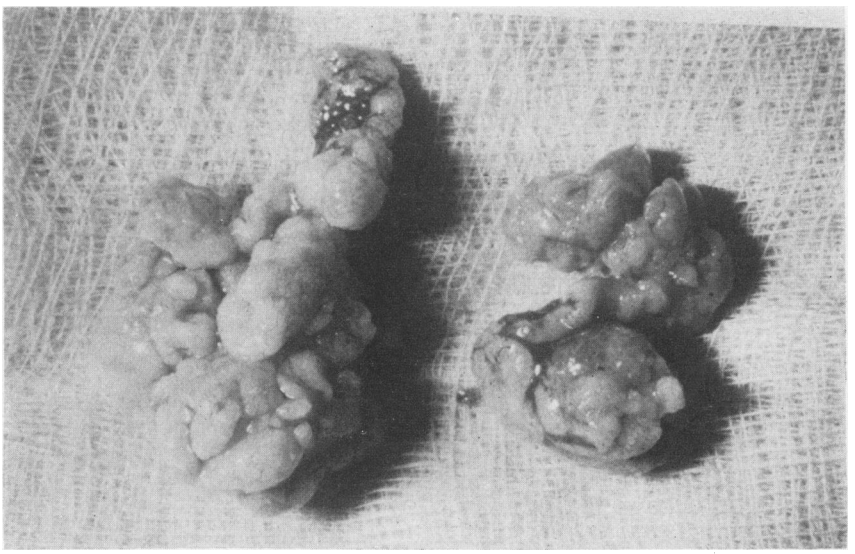

FIG. 4. Small bowel polyps.

malignancy remains uncertain. Carcinoma of the small bowel, the main site of polyposis, is rare and malignant change occurs more commonly in the colon, stomach or duodenum (Dozois *et al.*, 1969; Payson and Moumgis 1967; Williams and Knudsen, 1965). Female patients with PJS may develop ovarian tumours many of which are malignant (Dozois *et al.*, 1970).

Although the small bowel polyps were once considered pre-malignant adenomatous lesions, they are now regarded as benign hamartomas. The histological findings are characteristic; hamartomas have an excess of normal epithelium which covers strands of smooth muscle. Gastric and duodenal polyps also show hamartomatous features. Colonic polyps, however, are mainly adenomatous (Morson,

1962) and the gall bladder polyps in the present case fall into this category.

Routine oral cholecystography in patients with known PJS may show further cases of gall bladder polyposis. The occurrence of biliary colic in a patient with PJS should suggest the presence of gall bladder polyps probably associated with calculi.

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